Journal of Community Hospital Internal Medicine Perspectives

Volume 12 | Issue 5 Article 14

2022

Platypnea orthodeoxia for those who do better lying low

Rebecca DeBoer
Department of Medicine, Reading Hospital, rebecca.deboer@towerhealth.org

Tuoyo Mene-Afejuku
Department of Cardiology, Reading Hospital

Murtaza Sundhu
Department of Cardiology, Reading Hospital

Julian Diaz Fraga
Department of Cardiology, Reading Hospital

Recommended Citation

DeBoer, Rebecca; Mene-Afejuku, Tuoyo; Sundhu, Murtaza; and Fraga, Julian Diaz (2022) "Platypnea orthodeoxia for those who do better lying low," *Journal of Community Hospital Internal Medicine Perspectives*: Vol. 12: Iss. 5, Article 14.
DOI: 10.55729/2000-9666.1096
Available at: https://scholarlycommons.gbmc.org/jchimp/vol12/iss5/14

CASE REPORT

Platypnea Orthodeoxia for Those Who do Better Lying Low

Rebecca DeBoer [a,*], Tuoyo Mene-Afejuku [b], Murtaza Sundhu [b], Julian D. Fraga [b]

[a] Department of Medicine, Reading Hospital, Reading, PA, 19611, USA
[b] Department of Cardiology, Reading Hospital, Reading, PA, 19611, USA

Abstract

Platypnea orthodeoxia syndrome (POS) can be a confounding disease. Patients with POS may have already had an extensive and unrevealing evaluation for hypoxia. POS is the worsening of hypoxia when upright compared to supine. The underlying mechanism is a right to left shunt. While there are various causes of this, we focus on intracardiac shunt. The most common of these is patent foramen ovale (PFO). Once this is identified, closure of the PFO can lead to resolution of hypoxia.

Keywords: Platypnea orthodeoxia syndrome, Patent foramen ovale, Shunt, Hypoxia

1. Case presentation

A 78-year-old male with a history of obstructive sleep apnea, paroxysmal atrial fibrillation, and hypertension presented to our hospital after concerns for shortness of breath. He initially presented for an outpatient colonoscopy. The patient had worsening oxygen desaturation and so he was admitted.

He estimated that for the last 20 years he experienced shortness of breath. His breathing more acutely worsened over the past two years. He has been limited to solely walking around his property. Even walking 10 feet triggered his symptoms. About one month prior to current hospitalization, he was hospitalized for diverticulitis. He was sent home on 2 L oxygen.

Over the past two years he had an extensive work up for shortness of breath. He had pulmonary function tests and high-resolution computerized tomography (HRCT) scan of his chest. Both were unremarkable. He underwent cardiac catheterization and was found to have widely patent coronary arteries with mild calcification. He did not have underlying anemia. He was found to have obstructive sleep apnea and used a CPAP nightly. On this admission he also underwent computed tomography pulmonary angiography (CTA chest) that was unremarkable and there were no signs of pulmonary emboli.

During this hospitalization, it was noted that when he was sitting he was more hypoxic. When he was lying down his hypoxia improved. On our initial examination the patient was on high flow nasal cannula. His oxygen saturation was 92% when completely supine. With sitting and leaning forward he desaturated to 82%. With lying back down oxygenation improved to 91%. This was again repeated when the patient was titrated to 4 L nasal cannula. In supine position his oxygen saturation was 96%, sitting upright he desaturated to 86%, and returning to supine position he improved to 96%.

He underwent a transthoracic echocardiogram. Findings were reported as early positive agitated saline study consistent with patent foramen ovale or atrial septal defect. For better identification of intraatrial septal defect he underwent transesophageal echocardiogram. This showed right and left atria normal in size. Lipomatous hypertrophy of the intraatrial septum. The septum was aneurysmal. There was a PFO with significant right to left shunt by agitated saline injection. PFO tunnel measured

Received 4 February 2022; revised 24 May 2022; accepted 16 June 2022.
Available online 9 September 2022

* Corresponding author. Department of Medicine, Reading Hospital, Reading, PA, 19611, USA.
E-mail address: Rebecca.deboer@towerhealth.org (R. DeBoer).

https://doi.org/10.55729/2000-9666.1096

2 cm × 0.8 cm (Figs. 1–3). Review of CTA chest also did not show any definitive pulmonary arteriovenous malformations. The patient had right heart catheterization as well with post capillary wedge pressure (PCWP) 5 mmHg, right atrial (RA) pressure 3 mmHg, mean pulmonary artery (PA) pressure 14 mmHg. For oxygenation, high right atrial was 62%, mid right atrial 61%, low right atrial 63%.

The patient underwent PFO closure using a 35 mm Amplatzer PFO device. Post PFO closure patient's oxygenation was 95% supine and 95% in the sitting/upward position.

2. Discussion

Platypnea orthodeoxia is a rare and not fully understood syndrome.[1,2] There are three proposed mechanisms of platypnea orthodeoxia syndrome (POS). These are intracardiac shunt, intrapulmonary shunt such as pulmonary arteriovenous malformations, and ventilation perfusion mismatch.[1,2] The first is normally caused by a defect in the level of the atria.[2] Intracardiac shunts are caused by patent foramen ovale (PFO), atrial septal defect (ASD), or fenestrated atrial septa aneurysm.[2] In most cases the culprit is PFO.[2]

Of note, despite the prevalence of PFO, estimated to occur in 25% of the general population, only 2.2% of PFO led to POS.[1] Therefore, there are thought to be two other components that lead to right to left shunt.[2] First, some other functional abnormality within the heart occurs. For example, the patient may have aortic changes, persistent Eustachian valve, or lipomatous hypertrophy of the atrial septum.[2] Aortic changes include elongation, dilation, or aneurysm.[2] There are various reports of lipomatous hypertrophy of the atrial septum in patients with right to left shunt across a PFO.[3,4]

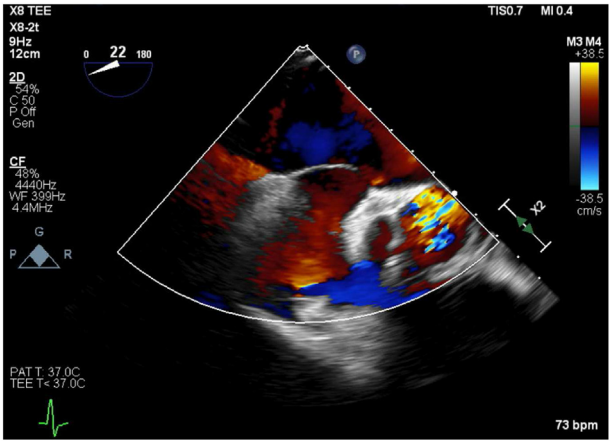

Fig. 1. Transesophageal echocardiography with doppler demonstrating patent foramen ovale with shunting into the left atrium.

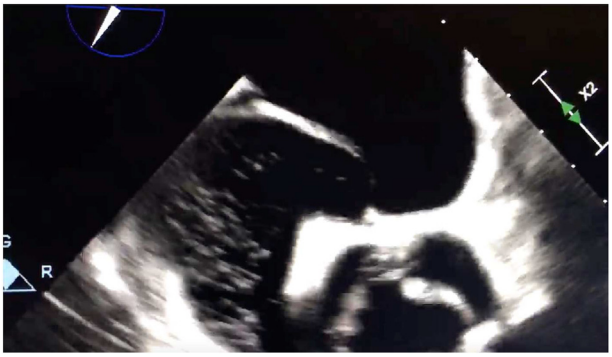

Fig. 2. Transesophageal echocardiography with bubble study beginning to demonstrate patent foramen ovale with shunting of bubbles into the left atrium.

Then there is some other precipitant that causes change in atrial flow, flow from the vena cava that drives blood through the PFO.[1] This precipitant may be time alone.[1] Both the PFO and other functional change are chronically present in these patients.[1] An aging heart with these predispositions may lead to the development of a right to left shunt.[2] In one review 38.5% of patients did not have any obvious trigger or underlying condition.[5] These patients were simply older than those with a noticeable predisposing factor.[5] In another review, 141 patients with right to left intraatrial shunt were all older than 50 years and on average in their seventh decade.[2]

There could also be a more acute trigger.[1] There could be a clear inciting event that causes the right to left shunt.[5] Potential triggers include recent pneumonectomy, underlying pulmonary pathology, ascending aortic dilation, or surgery.[5]

The diagnosis of POS is made with a thorough physical exam. On exam patients will have hypoxia in upright position and resolution or significant improvement in hypoxia with lying supine.[1] Postulated mechanisms for this change include that in

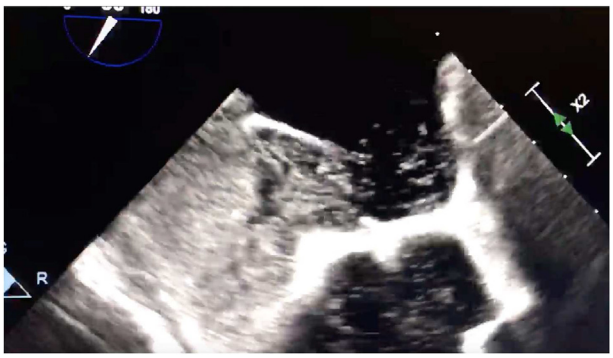

Fig. 3. Transesophageal echocardiography with bubble study demonstrating patent foramen ovale with shunting of bubbles into the left atrium.

sitting upright or standing the interatrial septum can be stretched increasing the size of the patent foramen ovale.[2] This augments blood flow through the opening.[2] Another theory is that sitting upright or standing shifts the interatrial spectrum horizontally, so the patent foramen ovale is in the direct path of blood flow coming to the right atrium from the inferior vena cava.[2]

Two literature reviews found that the improvement in oxygen with positional changes was on average between 10 and 12%.[1,5] Further investigation into the cause or type of shunt should start with transthoracic echocardiogram (TTE) with bubble study. When the bubbles are seen in the left atrium can also help categorize the type of shunt.[2] Bubbles in the left atrium within 3 beats signal an intracardiac shunt.[2] Transesophageal echocardiogram (TEE) can better visualize shunt if TTE is inconclusive.[1] These findings both on physical exam and imaging are enough for the diagnosis and evaluation of the syndrome.[1] Other tests can be performed. Pulmonary CT angiography can evaluate for intrapulmonary abnormality.[1] A right heart catheterization can be done as well. It will show normal pulmonary and right-sided pressures.[5] Usually the patient has already undergone extensive work up to rule out other etiologies of hypoxia.[1]

Treatment for POS secondary to PFO is closure of PFO.[1] This will immediately correct the hypoxia.[1]

Our patient had shortness of breath for years that had more recently worsened in the past two years. He had extensive workup that did not elucidate the cause of his shortness of breath and hypoxia. On physical exam he had a 10% drop in oxygenation when in the upright position. On evaluation with TEE he had significant right to left shunt through the PFO as well as lipomatous hypertrophy and aneurysm of the atrial septum. No obvious inciting event was discovered. He was in his 7th decade of life. Treatment did correct his hypoxia.

Our patient had very few comorbidities. This made shunt more of a plausible differential. However, other patients may have diseases such as heart failure or chronic obstructive pulmonary disease, confounding the diagnosis of shunt and specifically platypnea orthodeoxia syndrome.

3. Conclusion

Testing oxygenation supine and upright can be a valuable part of the workup for patients whose hypoxia is unexplained. This physical exam finding leads to the diagnosis of POS. To identify a shunt and also help determine where the shunt occurs, TTE and or TEE with positive bubble study should be performed. Right heart catheterization will reveal normal pulmonary and right-sided heart pressures. The importance in detection of PFO leading to POS is that treatment can resolve the hypoxia. This syndrome may have a predilection for older patients as time may further alter the already abnormal anatomy that causes POS. However, the exact mechanism for POS is not fully understood.

Disclaimers

We have no disclaimers.

Funding

We have no sources of support.

Declaration of competing interest

We have no conflicts of interest.

References

1. Blanche C, Noble S, Roffi M, et al. Platypnea-orthodeoxia syndrome in the elderly treated by percutaneous patent foramen ovale closure: a case series and literature review. *Eur J Intern Med*. 2013 Dec;24(8):813–817. https://doi.org/10.1016/j.ejim.2013.08.698. Epub 2013 Sep 3. PMID: 24007641.
2. Rodrigues P, Palma P, Sousa-Pereira L. Platypnea-orthodeoxia syndrome in review: defining a new disease? *Cardiology*. 2012; 123(1):15–23. https://doi.org/10.1159/000339872. Epub 2012 Aug 31. PMID: 22948714.
3. Sanikommu V, Lasorda D, Poornima I. Anatomical factors triggering platypnea-orthodeoxia in adults. *Clin Cardiol*. 2009 Nov;32(11):E55–E57. https://doi.org/10.1002/clc.20461. PMID: 19816867; PMCID: PMC6653461.
4. Shakur R, Ryding A, Timperley J, Becher H, Leeson P. Late emergence of platypnea orthodeoxia: chiari network and atrial septal hypertrophy demonstrated with transoesophageal echocardiography. *Eur J Echocardiogr*. 2008 Sep;9(5):694–696. https://doi.org/10.1093/ejechocard/jen015. Epub 2008 Feb 20. PMID: 18296402.
5. Shah AH, Osten M, Leventhal A, et al. Percutaneous intervention to treat platypnea-orthodeoxia syndrome: the toronto experience. *JACC Cardiovasc Interv*. 2016 Sep 26;9(18):1928–1938. https://doi.org/10.1016/j.jcin.2016.07.003. PMID: 27659570.